

# Anaesthetic Management of a Patient Having Ellis-Van Creveld Syndrome

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**Abstract:** *Ellis-Van Creveld Syndrome is rare Autosomal recessive condition characterized by short limb dwarfism, polydactyly, abnormal development of finger nails and congenital cardiac defects in 50% of the patients. The orofacial manifestation includes multiple gingivolabial fraenula, dental anomalies, hypodontia and malocclusion. The syndrome can be caused by mutation in EVC gene or EVC2 gene present in chromosome 4p16.*

**Keywords:** Ellis-Van-Creveld syndrome, polydactyly, Chondroectodermal dysplasia

## 1. Case Report

A 3 yr old female, weighing 11 kg came with complains of dental caries for 5 months and was posted for pulpectomy. She was the first child of non-consanguineous and normally developed parents. Her birth was full term normal vaginal delivery.

She had polydactyly in all 4 limbs, short limbs, conical teeth and multiple missing teeth.

On examination loud S1 and systolic murmur was heard in mitral region.

Her past medical history revealed that she was operated for AV canal repair and coronary sinus unroofing with left SVC ligation for severe mitral regurgitation at the age of 6 months.

She was a known case of Ellis-Van Creveld Syndrome and was posted for pulpectomy.



## 2. Preoperatively

Her vitals and labs were normal except for 2D ECHO which showed mild PAH, severe AR, grade 1 right AV Regurgitation and moderately dilated LA. Her airway assessment was normal.

She was on T. Enalapril 2.5 mg BD and T. Furosemide 2.5 mg BD.

Glycopyrrolate 0.004 mg/kg, Midazolam 0.05mg/kg and Inj. Fentanyl 2 mcg/kg was given as premedication.

## 3. Intraoperatively

She was induced with Inj Propofol 2mg/kg and Inj Atracurium 0.5 mg/kg and intubated with north pole uncuffed ET 4.5 and maintained on oxygen, Air, Sevoflurane and Atracurium. Inj Neostigmine 0.05mg/kg and Glycopyrrolate 0.008mg/kg was given and she was extubated.

Postoperatively she was sent to ward and discharged after one day.



